1. What orthopedic complications are seen in Ehlers Danlos Syndrome, and Hypermobility Spectrum Disorder patients?
   1. Patients with EDS and HSD have musculoskeletal complications that include joint subluxation and dislocations, tendon subluxation, and peripheral nerve problems. These problems tend to cause vague, intermittent pain that is often difficult to diagnose. Some joints wear prematurely, and can be painful long before there is radiographic evidence of a problem.
2. What is a dislocation? Subluxation?
   1. A “dislocation” is when the joint where two bones meet is displaced completely from its usual orientation. Dislocations of major joints like the hip, knee, or ankle, tend to be severe injuries, as they can damage nerves and arteries, in addition to ligaments. Dislocations of smaller joints, like the fingers, tend to be painful when dislocated but heal once put back in place and protected while they heal.
   2. “Subluxation” is partial dislocation, where the bones move more than they should, outside the normal path of movement, but the joint is not completely dislocated. Subluxation may or may not be painful. For example, inferior subluxation of the shoulder is common and is typically benign; cervical spine subluxation can cause spinal cord damage and can require urgent surgery.
3. When to manage dislocations at home and when to see a doctor?
   1. Joint subluxations, aside from those involving the spine, tend to be managed at home. True dislocations probably need to be evaluated by a physician, although it depends on which joint is dislocated. Spine instability can lead to soft tissue failure that can damage the spinal cord, and can require urgent surgery. Hip, knee and ankle dislocations are severe injuries and require emergency evaluation by a physician. Patella dislocations spontaneously resolve but may need surgery for persistent stability. Rib joints can pop in and out without causing major damage. Shoulders easily sublux, and can also dislocate in EDS/HSD patients. Repetitive shoulder dislocations can damage the cartilage and cause arthritis, and most of the time can be prevented with a specific exercise/rehabilitation program. Finger dislocations may need anesthesia to realign the joints, but finger splints are very effective to limit joint subluxation.
4. What imaging/tests are recommended to evaluate painful/unstable joints?
   1. Blood tests and imaging tests, including radiographs and MRIs, are frequently necessary to evaluate patients for painful joints, but in EDS/HSD patients they are typically normal, and not predictably helpful in evaluating for dislocations or subluxations, unless they include “stress views,” which do not exist for all joints. In addition to diagnostic tests, a physical exam by an educated provider knowledgeable in EDS/HSD is usually necessary, and more helpful.
5. What orthopedic procedures/surgeries are recommended for unstable joints?
   1. **Craniocervical instability** often requires a fusion for stability.
   2. **Cervical spine instability** with significant anterolisthesis or retrolisthesis can require fusion for protection of the spinal cord. Chronic instability can cause premature disk collapse and facet joint arthritis, and eventually nerve root compression.
   3. **Shoulder instability** usually responds to concentrated physical therapy, directed toward increasing the resting tone of the rotator cuff muscles. If physical therapy fails, Neer capsular shift surgery can result in stability. Labral tears are common in shoulder instability and generally respond to surgical repair, and require a MR arthrogram for diagnosis.
   4. Posterolateral rotatory **Instability of the elbow** is usually related to a traumatic event, and often requires surgical repair of the ulnar collateral ligament complex to stabilize the radiocapitellar joint.
   5. **Wrist instability** responds to capsular stabilization, but confirming the diagnosis can be extremely difficult, as few physicians are familiar with the type of wrist instability common in EDS patients, which is dynamic pseudo-dissociative carpal instability.
   6. **Thumb CMC joint** instability is common in EDS patients and responds to surgical stabilization with an Eaton-Littler ligament reconstruction using a tendon graft. Without stabilization, arthritis occurs at an earlier age.
   7. **Thumb MP joint** instability is common in EDS patients are can respond to ligament or capsule repair, but with a high failure rate. Fusion of the joint is more reliable, but increases the load above and below the fusion site.
   8. **Finger joint instability** responds reliably to splinting.
   9. **Tendon subluxation** in the hand can mimic joint instability complaints, and responds to surgical stabilization of the tendons.
   10. **Lumbar spine instability** can cause acute spinal cord compression, requiring immediate surgery for Cauda Equina Syndrome. Chronic instability can cause premature disk collapse and facet joint arthritis, and eventually nerve root compression.
   11. **Sacroiliac joint instability** usually responds to physical therapy, but if that fails the joint can be fused surgically.
   12. **Hip instability** often results in labral tears, which can be treated surgically, if diagnosed. MR arthrogram is needed to confirm the diagnosis. The results of labral tear surgery for the hip are better when allograft tissue is used instead of native tissue.
   13. **Knee instability** can respond to physical therapy, but often results in premature wear of the patella, and a higher incidence of ACL tears and medial meniscal tears, which respond to surgical repair.
   14. **Ankle instability** is common in EDS patients, and usually responds to orthotics to at least some degree. Ankle instability can be treated reliably with surgery in EDS/HSD patients when synthetic ligaments are used to augment the repair. Sinus tarsi stents (arthroereisis implants) are reliable for patients with pes planus and hindfoot valgus deformities. Straightening out the ankle can simultaneously improve knee, hip and back malalignment problems.
   15. **Metatarsalgia** is common in loose jointed patients, and often responds to orthotics.
   16. **Bunions** are common in loose jointed patients, and often respond to orthotics. Surgery is an option, but the failure and recurrence rates are higher in EDS/HSD patients.
6. Which procedures and surgeries should be avoided?
   1. Generally speaking, non-surgical treatment is preferable. When non-operative measures fail or are ineffective, surgery may be the only option for reliable relief from painful joint or tendon instability or peripheral nerve compression, but should be performed by a surgeon familiar with EDS, in the context of overall management by a team of knowledgeable providers. The problems with surgery start before the operation, where the diagnosis may not be clear, or the diagnosis may be incomplete. Complications from surgery can easily negate the benefit, and surgical intervention should be approached cautiously by all involved. Large incisions should be avoided. Fusions may be necessary, but can increase the load above and below the fusion site, and can lead to other problems. Soft tissue procedures involving repair of native tissues tend to have a higher failure rate, and newer techniques involving allografts and ligament augmentation devices seem to have a higher success rate. Patients should probably avoid surgery by providers who are early in their careers or who do not have specific experience with EDS patients.
7. When should surgery to stabilize joints be done?
   1. When non-operative measures fail, there are no other options, and the potential results of surgery outweigh the risks involved. The thumb CMC joint should be stabilized early, to prevent or at least delay arthritis.
8. Is fusing a joint that won’t stop dislocating or subluxing ever recommended?
   1. Yes. Sacroiliac joints, thumb CMC and MP joints, ankle and cervical spine joints can be fused. Fusing the shoulder, elbow, hip and knee cause significant disability. For some joints, such as the spine and the thumb, fusing a joint increases the load at the adjacent joints in a system that has already failed, and fixing one problem can create two more problems over time.
9. Are PRP/prolotherapy/stem cell injections helpful for EDS?
   1. The role of these treatments is unclear at the moment, but under certain circumstances, these treatments can be extremely helpful. The results can be unpredictable, but the risks tend to be smaller than surgical intervention, but the procedures are not typically covered by insurance. Stay tuned for further developments.
10. Are braces/splints helpful for joint instability?
    1. Braces can be extremely helpful for joint instability, particularly when used in conjunction with a supervised physical therapy program directed by a knowledgeable provider.
11. Do those with a type of EDS or HSD lose hypermobility as they age?
    1. Joints tend to get looser with age, not tighter, although as joints become arthritic they tend to become stiffer, such as the neck or the thumb CMC joint. The muscles tend to become tighter, which can result in a confusing physical exam.
12. Do those with a type of EDS or HSD gain joint stability as they age?
    1. Joints that become arthritic from excess motion tend to become stiffer with age. Non-arthritic joints tend to remain loose, but the muscles can become tight as people adapt to the arthritis to avoid pain, which can be confusing.
13. When do those with a type of EDS or HSD develop osteoarthritis? What are the symptoms and signs? How is it diagnosed and treated?
    1. Everyone develops arthritis in their joints if they live long enough. Joint wear comes from a combination of load and motion, and hypermobile patients have excess motion, and the joints tend to wear prematurely. Hypermobile joints tend to be sore for a long time before there is actual arthritis, ie damage to the joint surface. On a radiograph, this would present as joint space narrowing (from loss of cartilage; cartilage does not show on x-rays), sclerosis (increased bone density) and osteophytes (bone spurs).
    2. In hypermobile patients, the first joint that tends to actually become arthritic is the patellofemoral joint. This causes anterior knee pain when descending stairs. The second joint to wear out is the thumb CMC joint. The problems with spine arthritis progress with time, but by age 50 tend to make patients seek medical attention for that arthritis.
14. Are those with a type of EDS/HSD more prone to fractures?
    1. In my experience, fractures are less common in hypermobile patients, because the ligaments fail first, and that tends to protect the bones. However, EDS/HSD patients often fall from ankle, knee, or hip instability, and can break their wrist or injure their thumb when they fall.
    2. Some of the rarer types of EDS are associated with osteopenia, and those with spEDS are prone to spontaneous fractures.
15. Are spinal curve abnormalities seen in types of EDS and HSD?
    1. Scoliosis
    2. Kyphosis
    3. Cervical spine straightening (“loss of lordosis”)
16. Aside from the above, are there other often asked questions by patients, or observations you make in the clinic and that you are able to share and answer here for additional information?
    1. It is likely that patients with loose joints have soft nerves, and nerve complaints are much more common in this patient population. Treatment of the nerve complaints requires comprehensive understanding of the anatomy and pathophysiology of the nervous system, and the effects of poor posture on the nerves in the upper extremity.

**How safe are steroid injections?**

* 1. Some EDS patients refuse steroid injections because the injections can damage ligaments. It is true that steroid injections into ligaments should be avoided. However, there are different kinds steroids that can be injected, and some are less damaging than others. Triamcinolone (Kenalog) tends to cause skin depigmentation and fat atrophy if injected near skin, and particularly weakens ligament tissue. Betamethasone (Celestone) is much less damaging, and can be injected near joints (not into ligaments) and into tendon sheaths without damage. If a patient needs more that 2 injections in one place, they probably need a different treatment. Multiple injections in the same area should be avoided. Steroid injections can be considered safe in EDS/HSD patients when given by skilled, knowledgeable providers who understand these issues.

**What about local anesthesia?**

* 1. Patients with EDS/HSD are sometimes resistant to the usual local anesthetic agents that doctors use for injections, specifically lidocaine (Xylocaine) and bupivacaine (Marcaine). In addition, these medications are known to be specifically cytotoxic to chondrocytes at normal concentrations. That is to say that these medications are poisonous to cartilage cells, and generally speaking should not be injected into joints unless diluted. Ropivacaine (Naropin) is much safer to inject into joints. For injections other than joints, mepivacaine (Polocaine, Carbocaine) seems to work more reliably in EDS/HSD than lidocaine or bupivacaine.

**What about the fluoroquinolone class of antibiotics?**

* 1. Fluoroquinolones are powerful antibiotics with excellent tissue penetration that are used routinely for oral, respiratory, soft tissue and urinary tract bacterial infections. Name brands include ciprofloxacin (Cipro), levofloxacin (Levaquin), moxifloxacin (Avelox), norfloxacin (Noroxin), ofloxacin (Floxin). These medications can have serious side effects that can be more of a problem for EDS/HSD patients. Specifically, they can cause nerve damage called peripheral neuropathy, and there is an increased risk for tendon rupture/avulsion, particularly of the Achilles tendon, and should be avoided if possible.

The Ehlers Danlos Society

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